ICD – First

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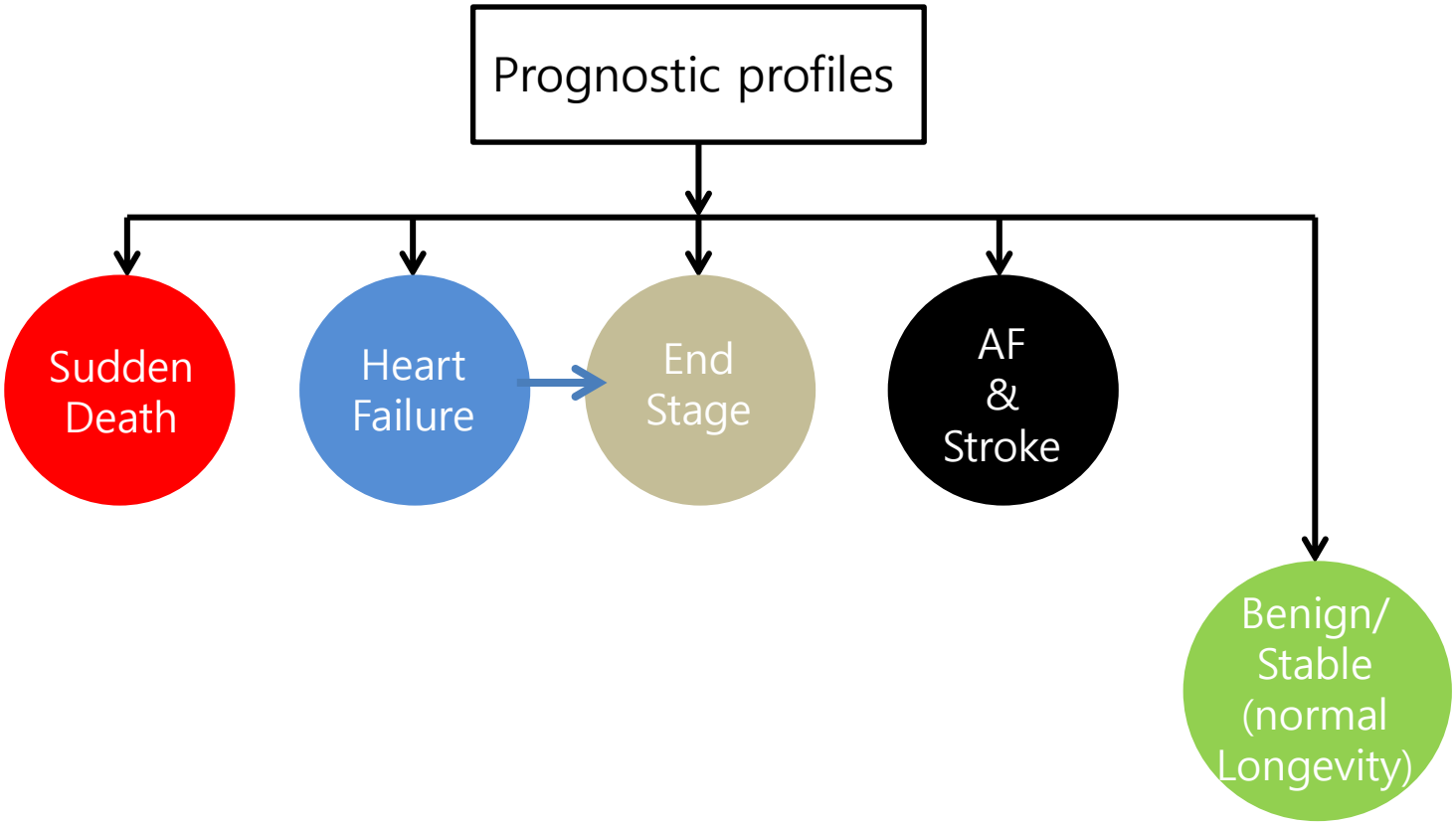
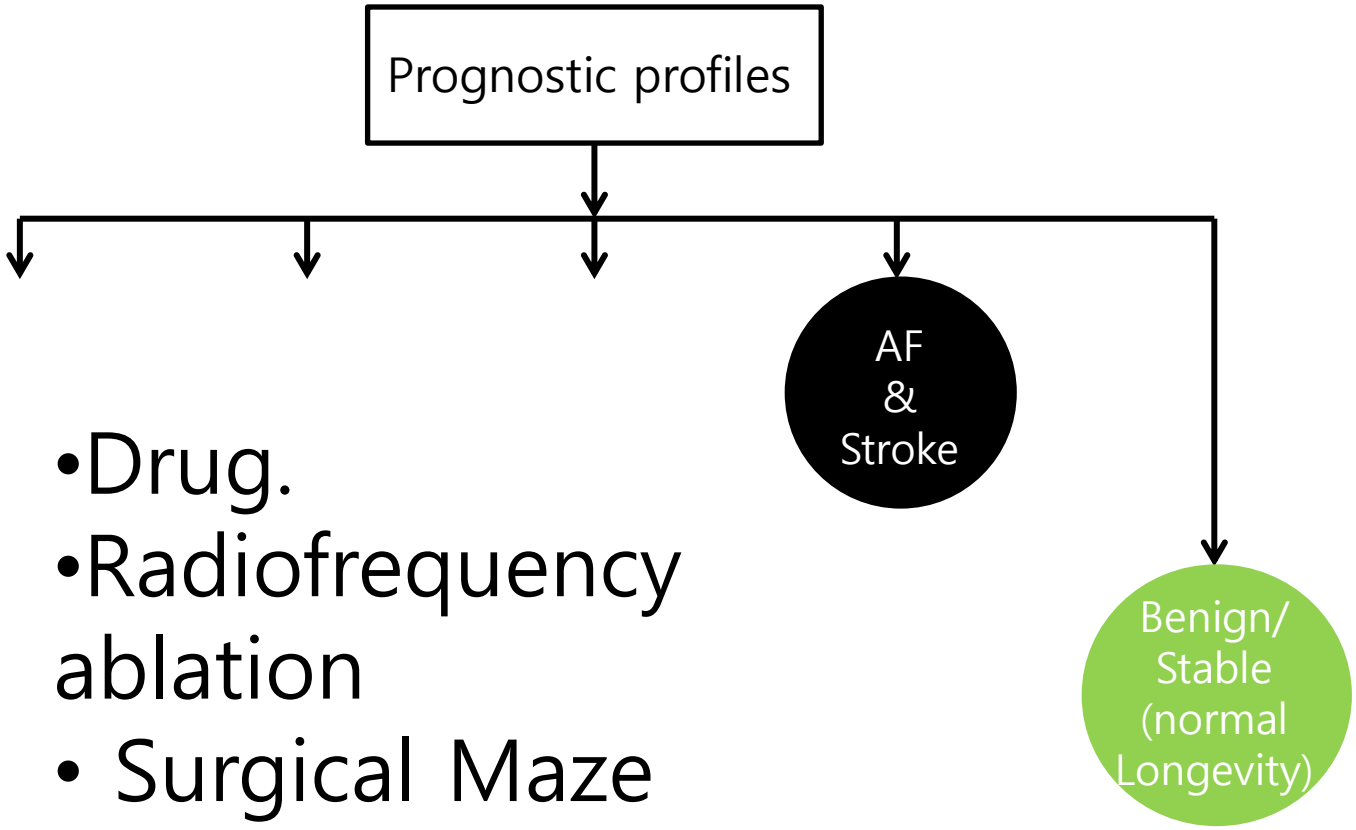
HCM

- HCM is heterogenous cardiac disease with a diverse clinical presentation and course in all age groups.
- HCMP is the most common cause of SCD in young people.
- SCD usually occur in previously healthy individuals without Sx or as the initial clinical manifestation of the disease.
- Annual SCD rate is <1% among HCM

Natural Hx and Tx Intervention

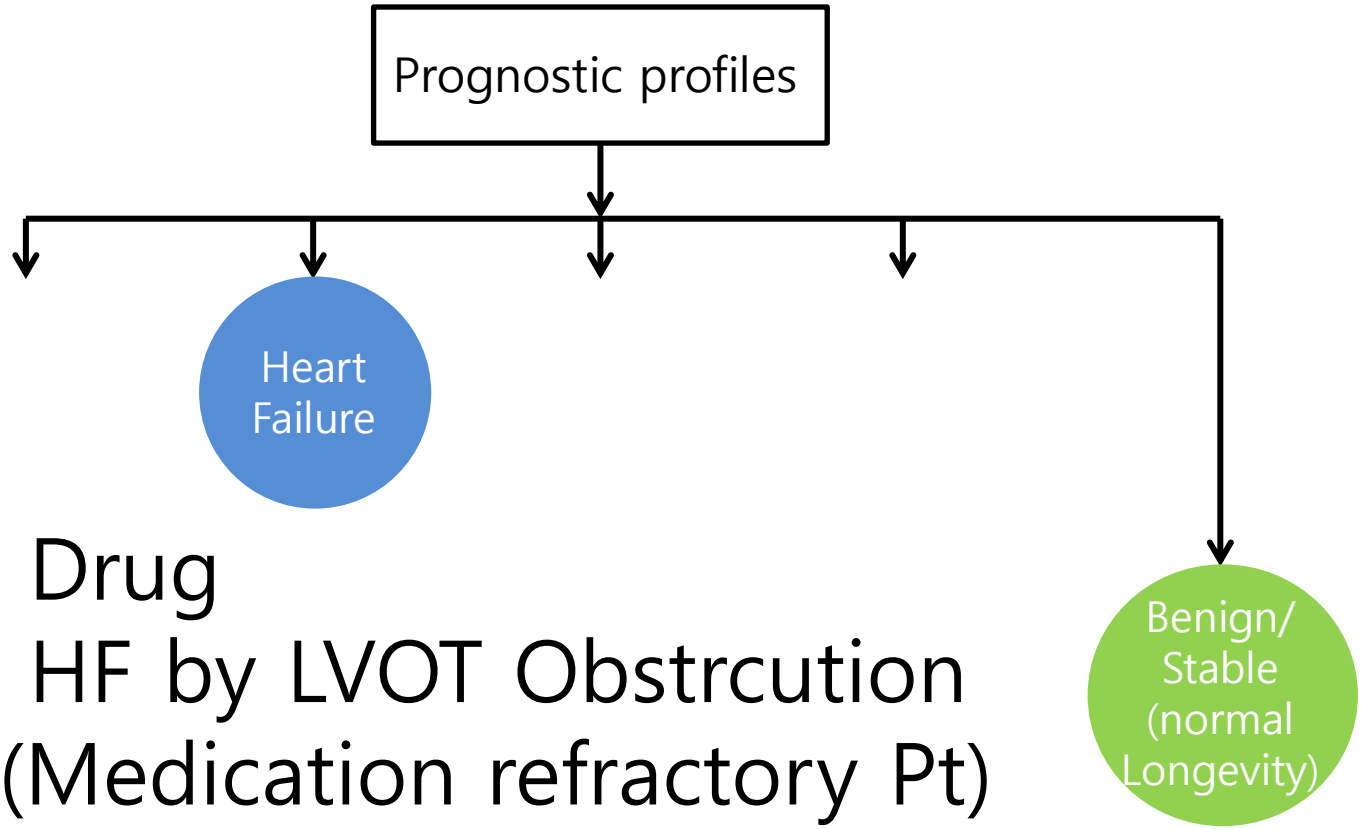


Natural Hx and Tx Intervention



- Drug.
- Radiofrequency ablation
- Surgical Maze

Natural Hx and Tx Intervention



- Drug
- HF by LVOT Obstrcution (Medication refractory Pt)

-Myectomy

-Alcohol septal ablation

Definition of Dynamic LVOT obstuction

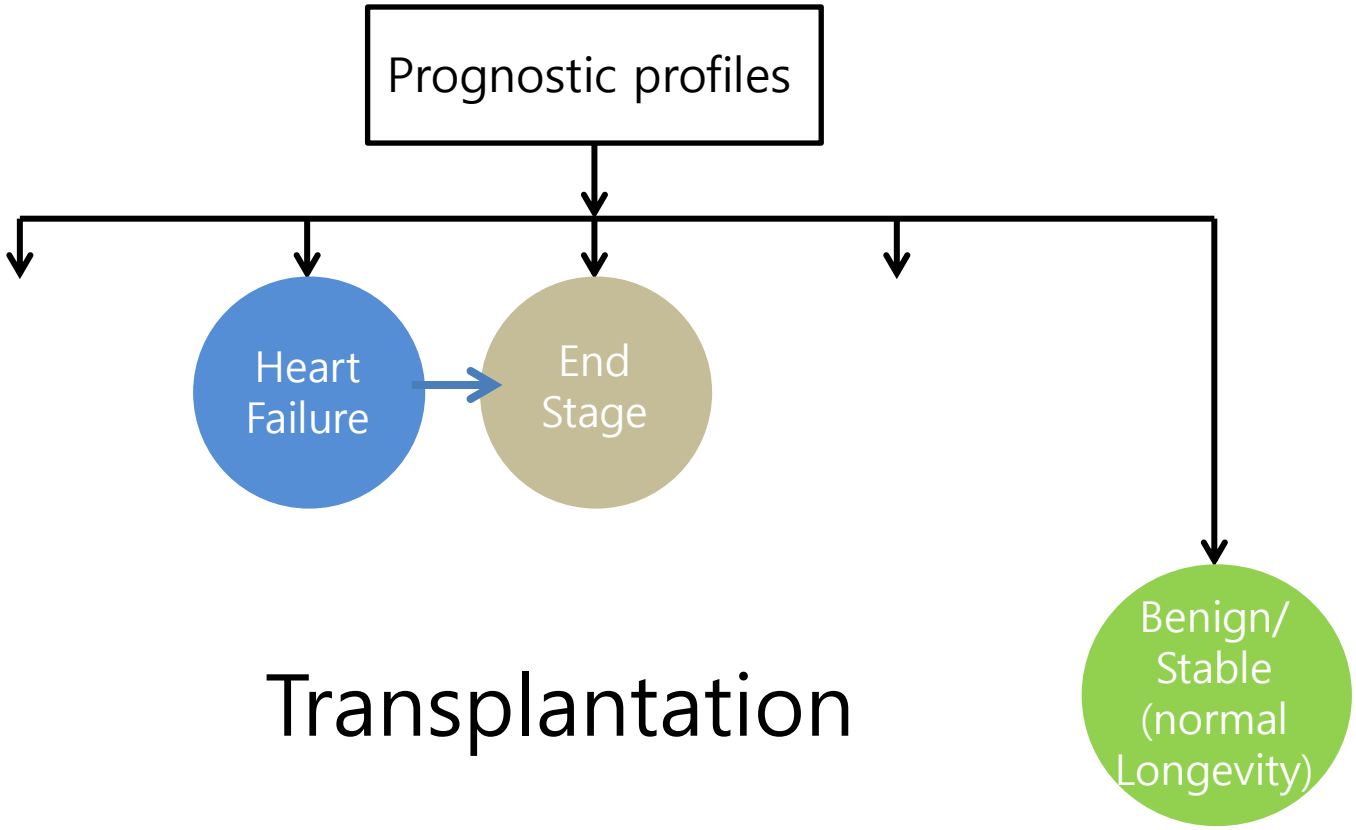
Hemodynamic State	Conditions	Outflow Gradient*
Basal obstruction	Rest	≥30 mm Hg†
Nonobstructive	Rest	<30 mm Hg
	Physiologically provoked	<30 mm Hg
Labile obstruction	Rest	<30 mm Hg†
	Physiologically provoked	≥30 mm Hg†

*Either the peak instantaneous continuous wave Doppler gradient or the peak-to-peak cardiac catheterization gradient, which are equivalent in hypertrophic cardiomyopathy.[73,74]

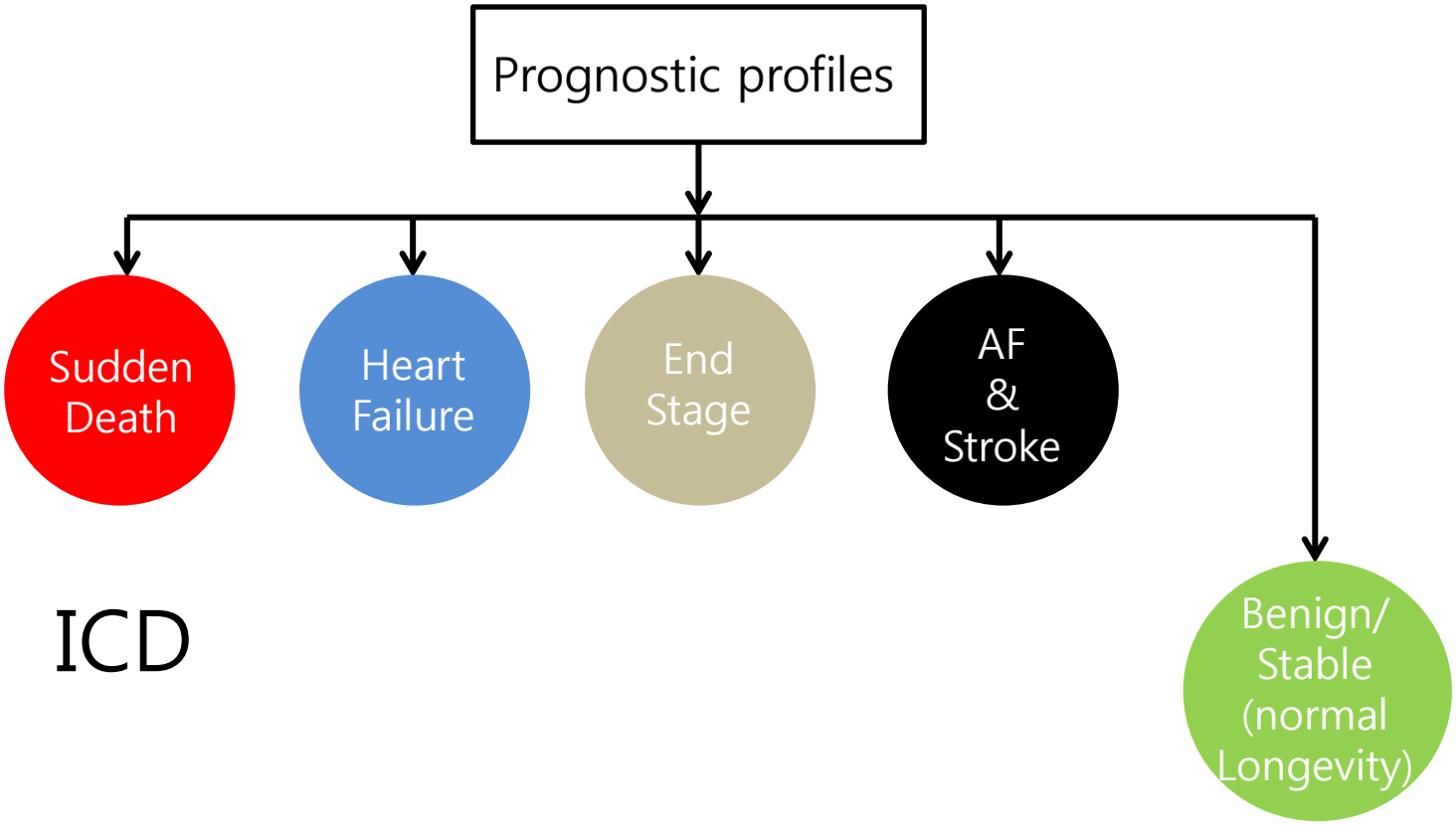
†Gradients ≥50 mm Hg either at rest or with provocation are considered the threshold for septal reduction therapy in severely symptomatic patients.

2011 ACCF/AHA guideline. Circulation 2011;124:e783-e831

Natural Hx and Tx Intervention



Natural Hx and Tx Intervention



ICD

Risk factor stratification for SCD

Identification of high risk patients and effort at prevention of SCD represent important clinical challenges in HCM.

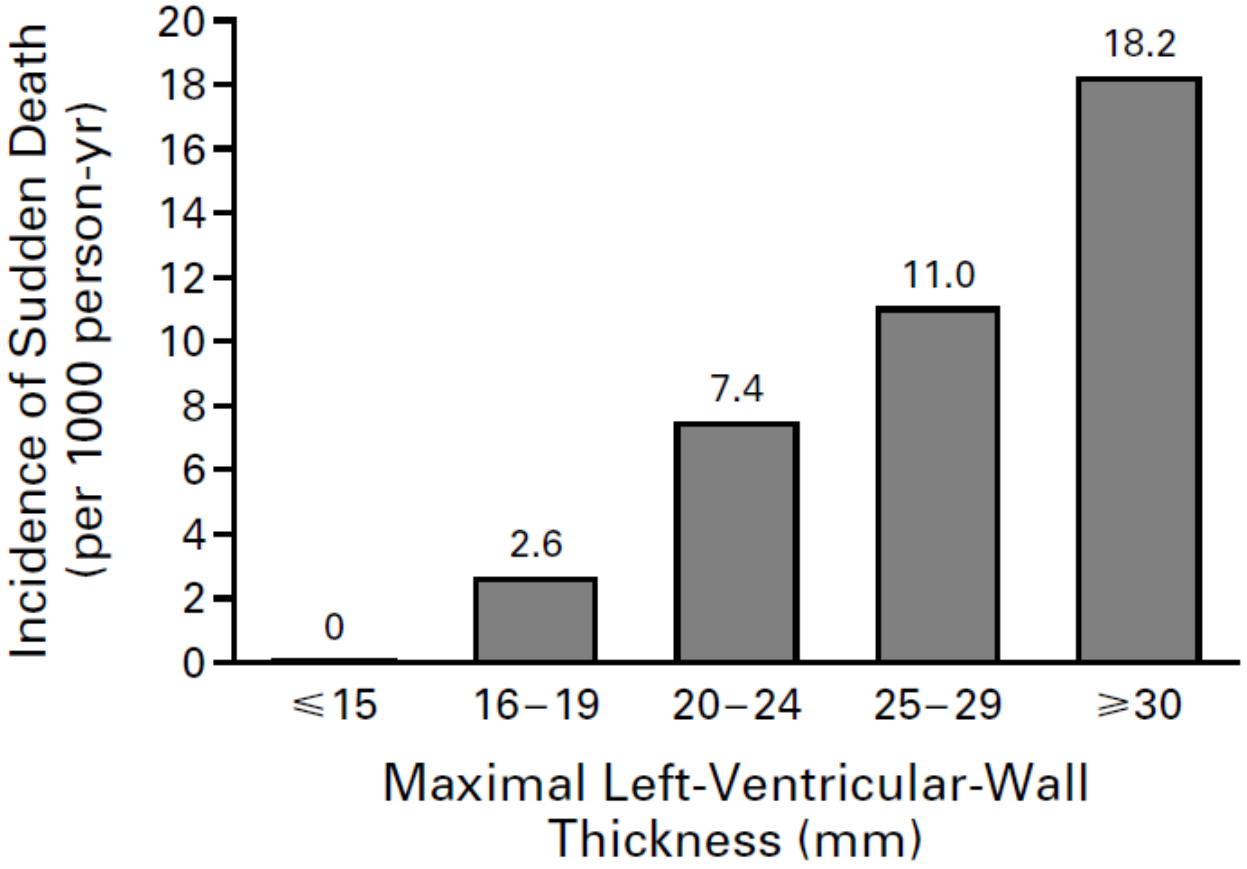
Risk Factors for SCD in HCM

Major	Possible in Individual Patients
Cardiac arrest (ventricular fibrillation)	Atrial fibrillation
Spontaneous sustained ventricular tachycardia	Myocardial ischemia
Family history of premature sudden death	LV outflow obstruction
Unexplained syncope	High-risk mutation
LV thickness greater than or equal to 30 mm	Intense (competitive) physical exertion
Abnormal exercise blood pressure	
Nonsustained ventricular tachycardia (Holter)	

ACC/ESC 2003 Clinical Expert Consensus on HCM. *JACC. 2003;9:1687-713*

Relation between LVH and SCD

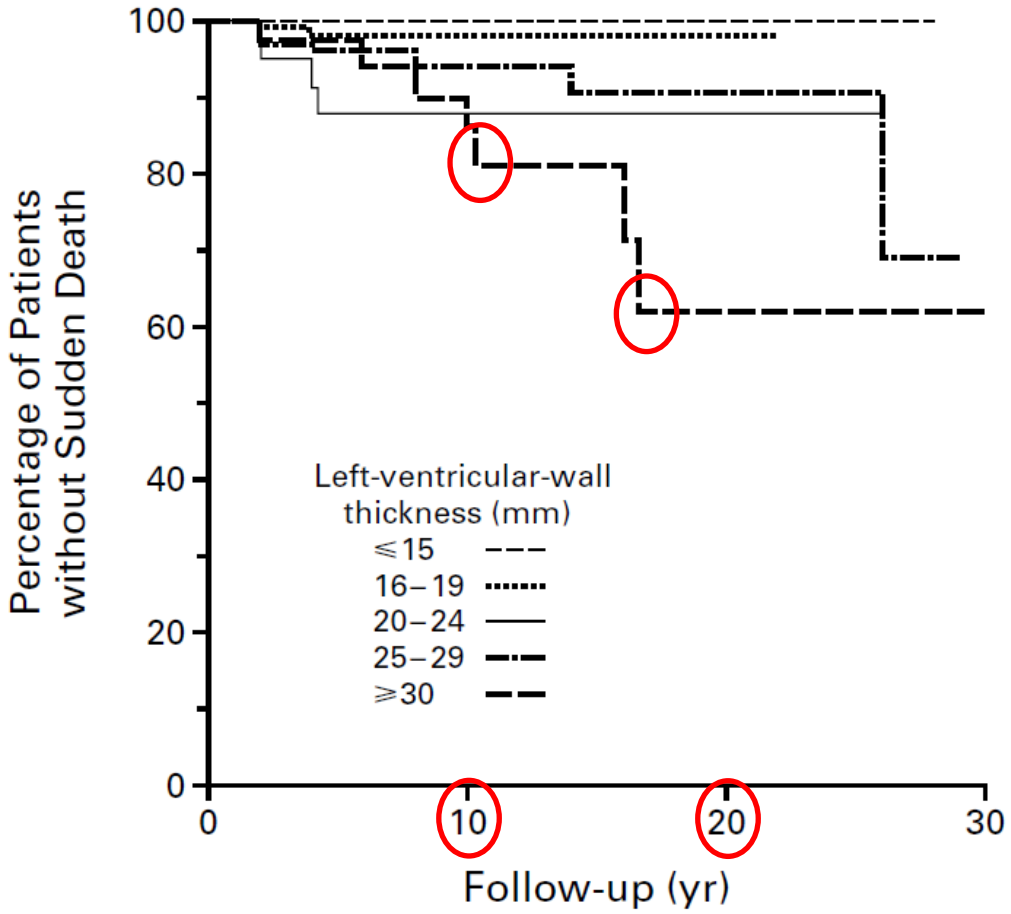
Risk of Sudden Death in 480 Patients with HCMP



N Engl J Med 2000;342:1778-85

Relation between LVH and SCD

Kaplan-Meier Estimate of the proportions of patient without SCD

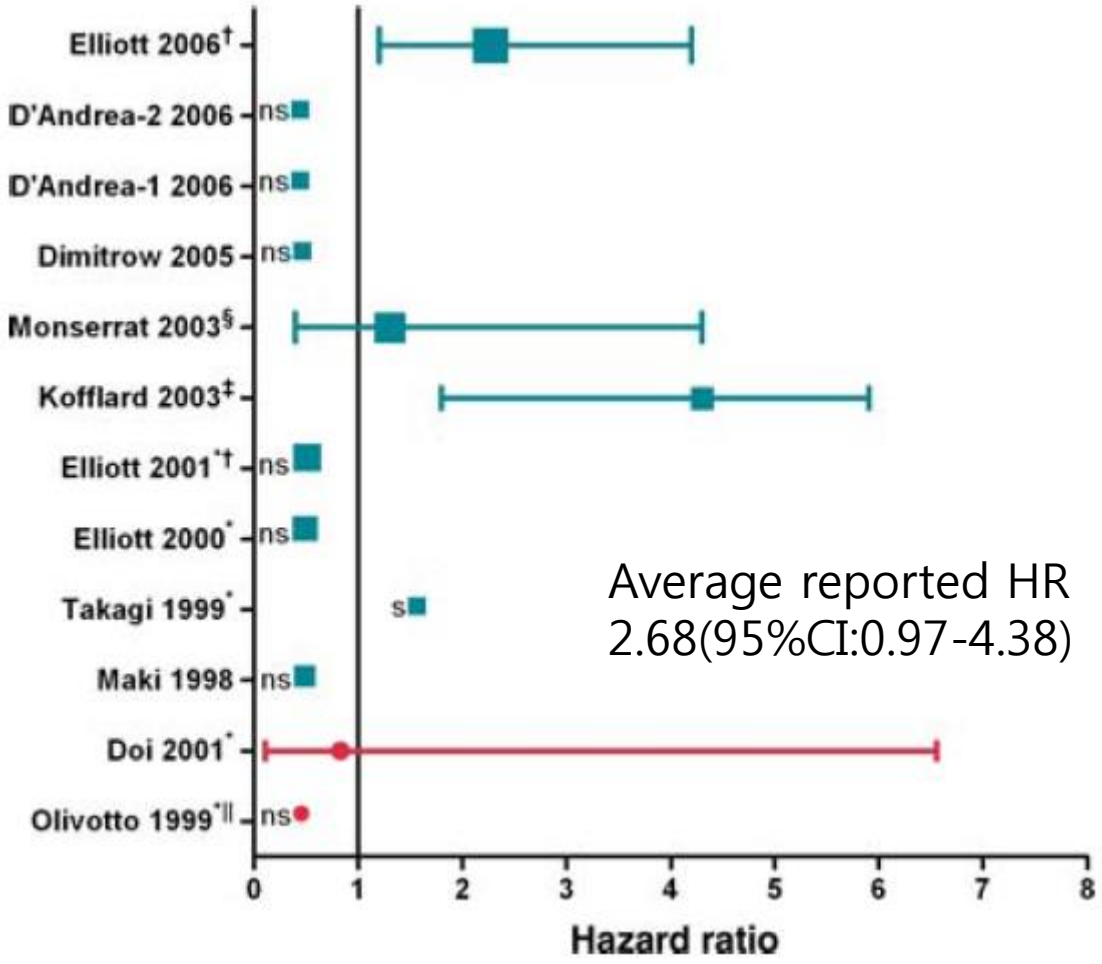


N Engl J Med 2000;342:1778-85

Syncope in HCM

- Syncope is a complex entity since several mechanism may be responsible for this Sx.
- Multiple cause of Syncope
  - SVT,
  - Bradyarrhythmia,
  - Ventricular arrhythmia,
  - Abnormal vascular response
  - Exercise-related LVOTO
  - Ischemia
  - Neurally mediated syncope
  - Orthostatic hypotension

Relation between Unexplained Syncope and SCD

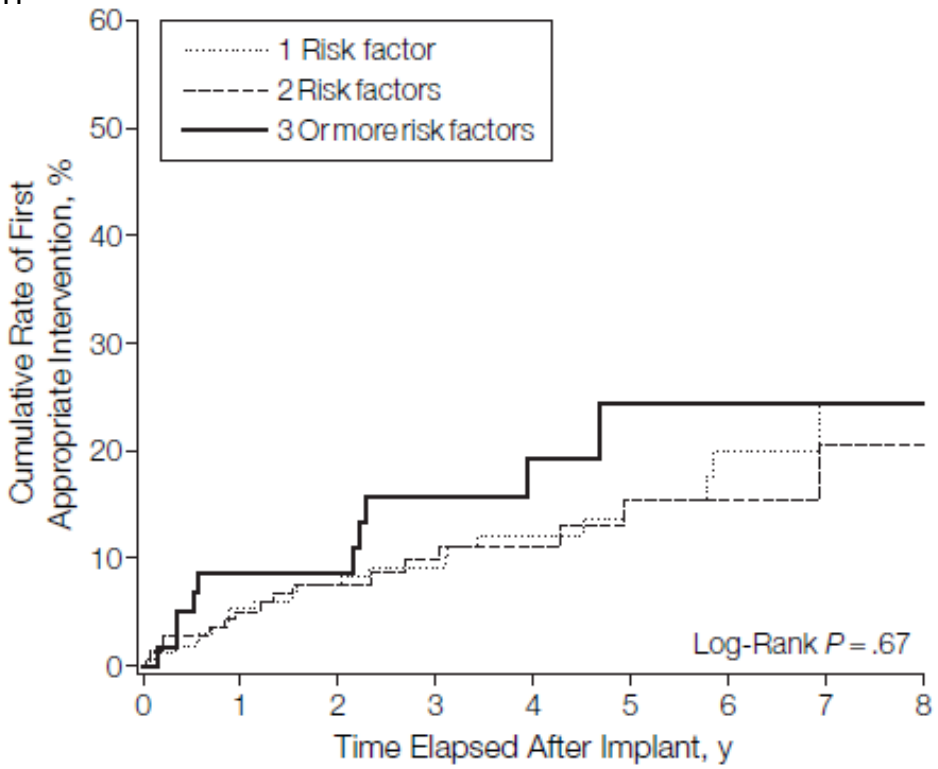


Average reported HR
2.68(95%CI:0.97-4.38)

Europace 2010;12:313-321

Relation between number of risk factors and SCD

Cumulative rate for first ICD intervention in pt with 1,2, or 3 or more risk factors who had received devices for primary prevention



No. at risk									
1 Risk factor	173	150	119	98	70	48	31	18	16
2 Risk factors	143	123	95	71	53	34	28	16	6
3 Or more risk factors	59	52	38	32	23	11	9	8	6

JAMA 2007;298(4):405-412

SCD in HCM

- SCD in HCM is caused mainly by Ventricular arrhythmia that can be effectively treated by ICD

JAMA 2007;298:405-12

Mechanism of SCD

- Substrate : anatomical substrate

  GDE on CMR: myocardial fibrosis and disarray
- Trigger
  - Myocardial ischemia
  - LVOTO
  - Change in vascular architecture
  - Atrial fibrillation/enhanced AVN conduction

Mechanism of Myectomy for SCD Prevention in HCM

- Removal of LVH
  - Removal of anatomical substrate
  - Reduction of myocardial oxygen demand, coronary vascular resistance and capillary density.
- Reduction of LVOTO
  - Prevention severe reduction in CO leading to electromechanical dissociation
  - Prevention ventricular arrhythmia through myocardial ischemia

Percentage of Risk factors

- **Almost 5% of pt without any risk factors experience sudden cardiac death.**

J Am Coll Cardiol 2000;36:2212-2218

Mechanism of SCD

- Substrate : anatomical substrate

  GDE on CMR: myocardial fibrosis and disarray
- Trigger
  - Myocardial ischemia
  - LVOTO
  - Change in vascular architecture
  - Atrial fibrillation/enhanced AVN conduction

- **Until now, We don't know exact mechanism**

Prevention of SCD: ICD

- ICD is the gold standard treatment for both the primary and secondary prevention of SCD in HCM.
- SCD is rare in ICD recipients and they receive appropriate device therapies that terminate ventricular arrhythmias.

Efficacy of ICD in HCM Pt with high risk for SCD

Study	Patient number, setting	Implant dates	Mean age at implant (years)	NYHA I/II	Age ≤16 years	Male sex	Primary prevention	Mean follow-up (years)
Primo *et al*[9]	13, 2 centres	NA	48±13	NA	Yes	62%	15%	2.2
Maron *et al*[7]	128, 19 centres	1984–1998	40±16	86%	Yes	69%	66%	3.1
Begley *et al*[4]	132, 1 centre	1987–2001	34±17	NA	Yes	61%	64%	4.8
Jayatilleke *et al*[5]	22, 1 centre	1997–2003	NA	NA	NA	NA	82%	2.9
Marin *et al*[12]¶	45, 3 centres	2000–2005	43±20	91%	Yes	62%	60%	2.5
Woo *et al*[10]	61, 1 centre	1996–2003	46±18	NA	Yes	66%	82%	3.3
Kaski *et al*[11]	22, 1 centre	1993–2006	14	84%	Yes	59%	77%	1.7
Maron *et al*[8] **	506, 42 centres	1986–2003	42±17	87%	Yes	64%	76%	3.7
Lin *et al*[6]	181, 1 centre	1988–2005	44±17	NA	Yes	62%	86%	4.9
Syska *et al*[13]	104, 1 centre	1996–2006	36±17	95%	NA	45%	75%	4.6
The Heart Hospital	334, 1 centre	1992–2009	42±14	92%	No	62%	92%	3.8

Heart 2012;98:116-125

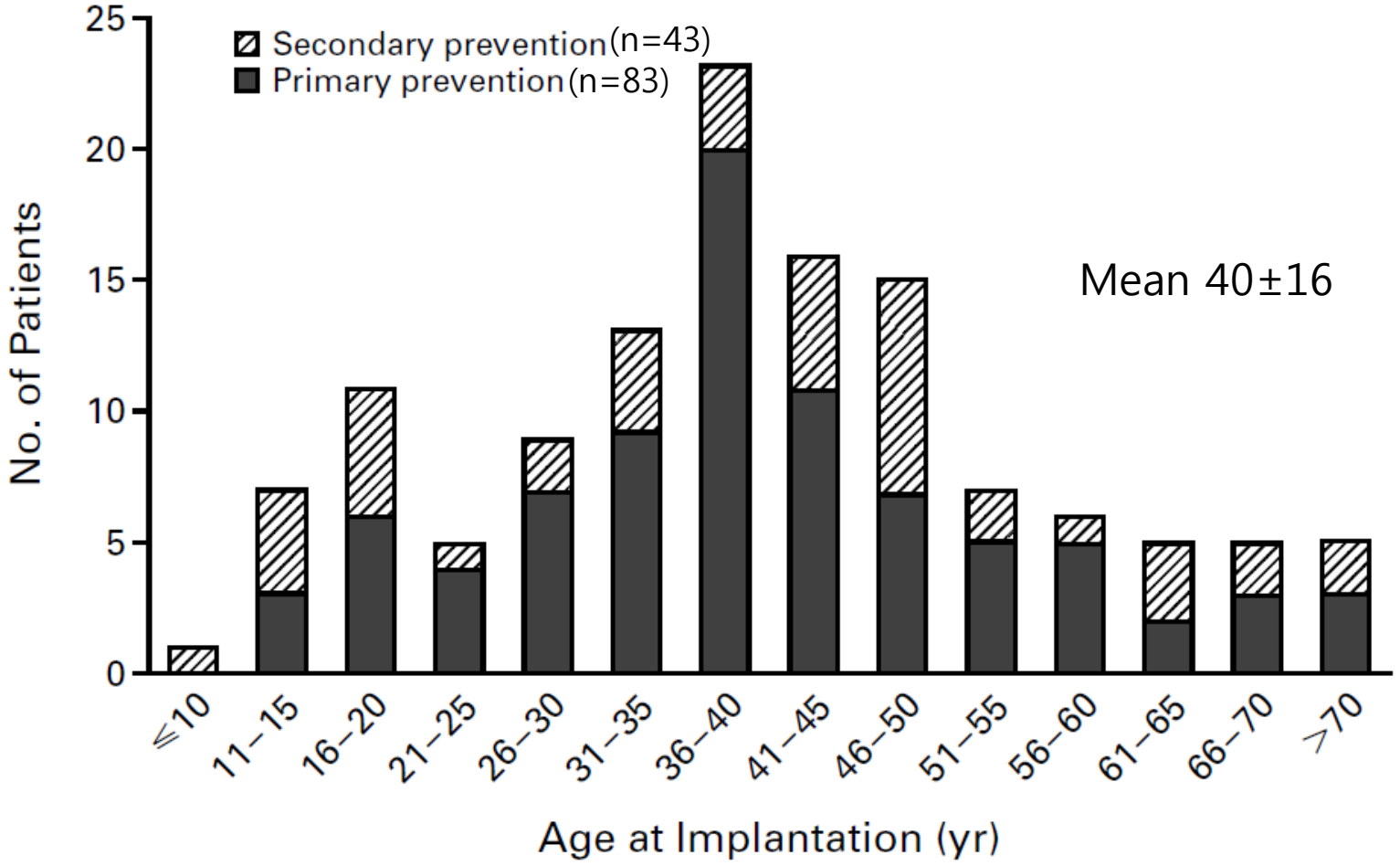
Efficacy of ICD in HCM Pt with high risk for SCD

Study	Appropriate shock rates*	Primary prevention appropriate shock rates	Secondary prevention appropriate shock rates	Inappropriate shocks (% of patients)	Implant complications† (% of patients)	Cardiovascular mortality‡	
Primo *et al*[9]	21% at 4 years	NA	NA	23	NA	0	
✓ Maron *et al*[7]	7%/year	5%/year	11%/year	25	14	NA§	
Begley *et al*[4]	25% at 5 years	16% in 5-years	36% in 5-years	23	10	3% deaths, 0.8% transplants	
Jayatilleke *et al*[5]	11%/year	10%/year	17%/year	9	5	NA	
Marin *et al*[12]¶	7%/year	1.6%/year	11.1%/year	27	2.2	4% deaths	
Woo *et al*[10]	4%/year	NA	NA	33	13#	2% deaths, 2% transplants	1
Kaski *et al*[11]	13%/year, 20% at 5 years	4.1%/year	71%/year	18	18	0	
Maron *et al*[8] **	5.5%/year, 23% at 5 years	3.6%/year	10.6%/year	27	12	4% deaths, 2% transplants	1
Lin *et al*[6]	4%/year	NA	NA	23	26	4% deaths, 2% transplants	
Syska *et al*[13]	5.6%/year	4.0%/year	7.9%/year	34	24	3% deaths, 1% transplant	1
The Heart Hospital	2.3%/year, 13% at 5 years	2.0%/year	4.3%/year	16	18	3% deaths, 3% transplants	0

SCD is rare

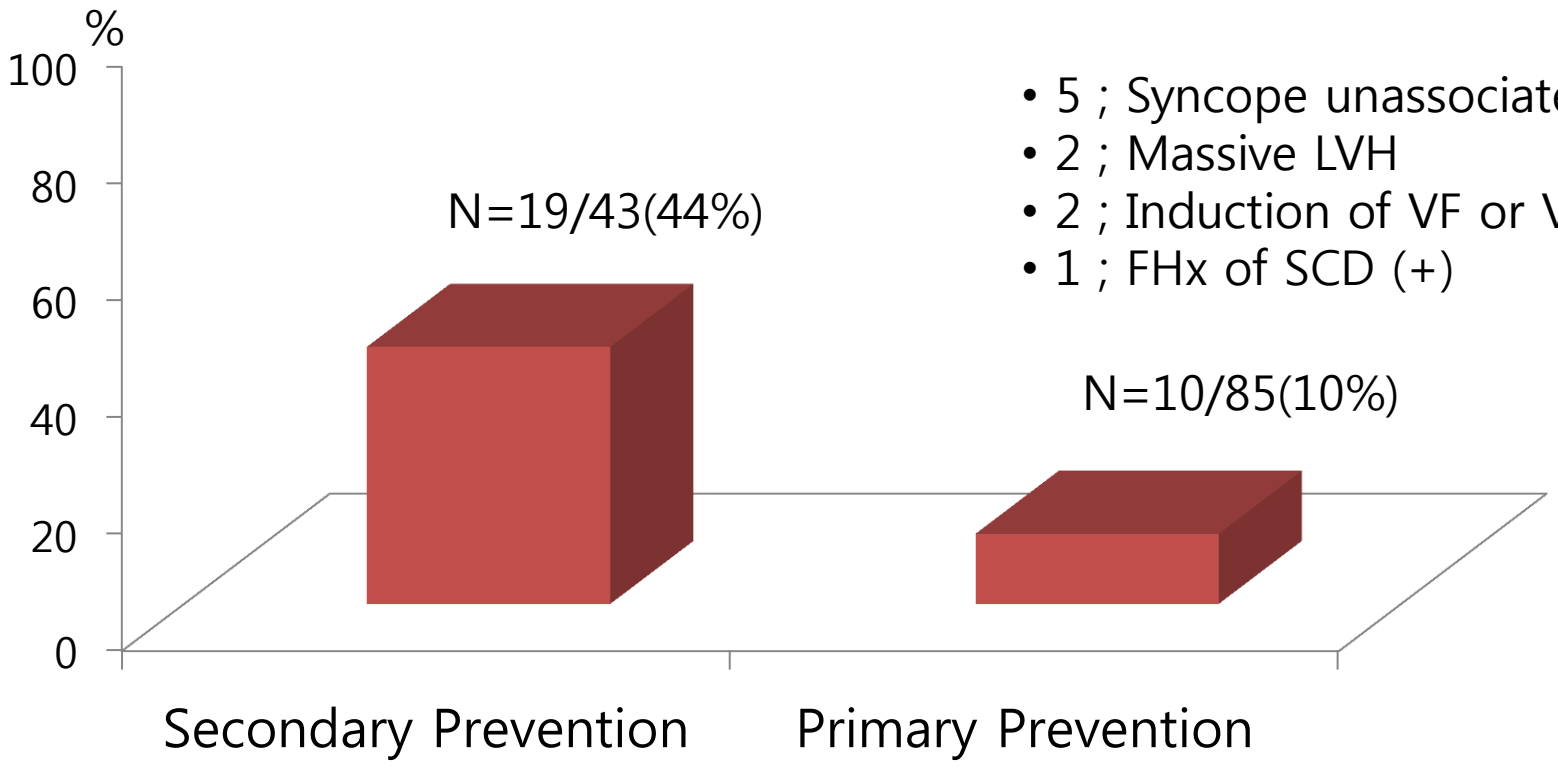
Heart 2012;98:116-125

Population of Patients



N Engl J Med 2000;342:365-73

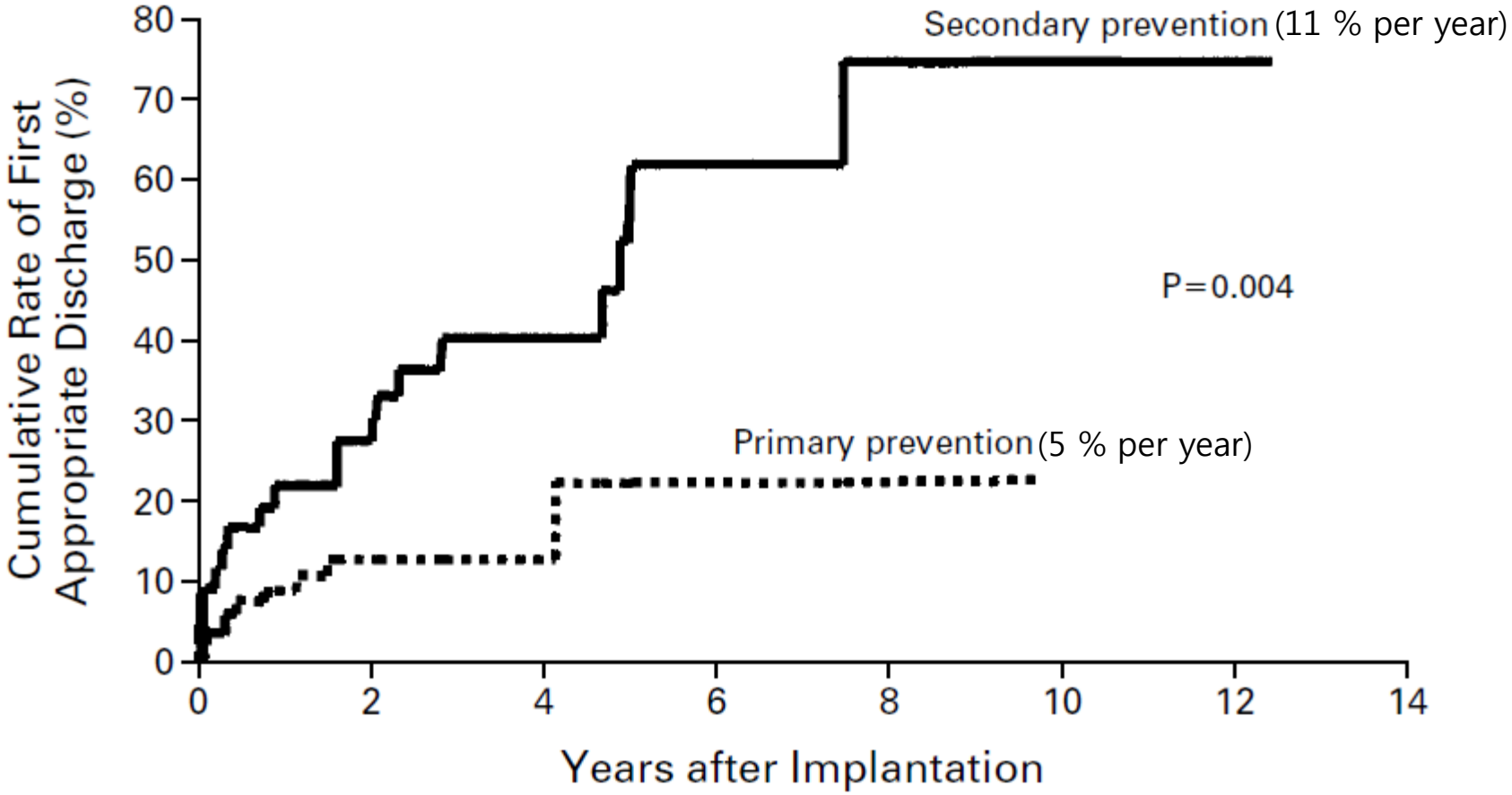
Appropriate Discharges



- 5 ; Syncope unassociated with VT
- 2 ; Massive LVH
- 2 ; Induction of VF or VT on EPS
- 1 ; FHx of SCD (+)

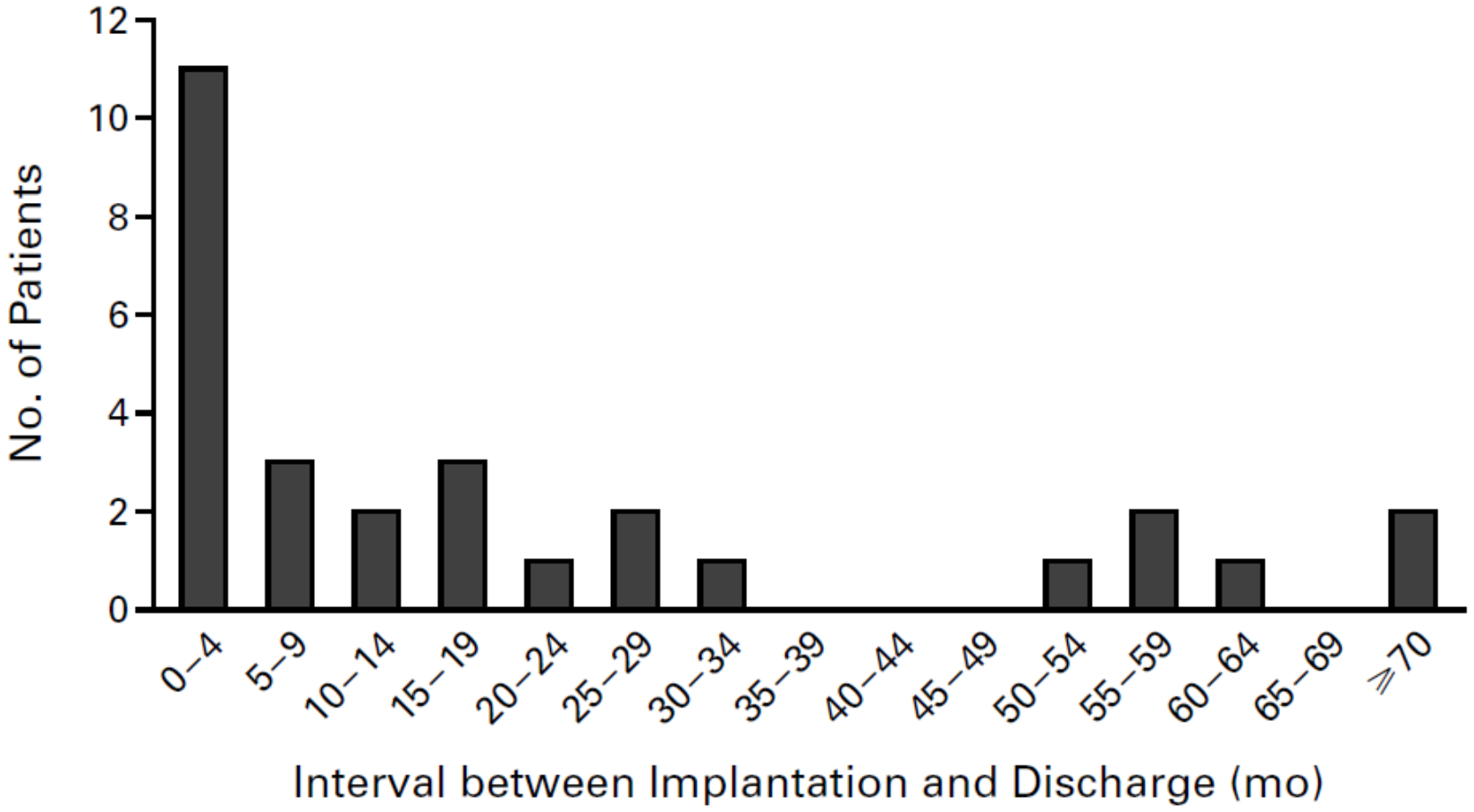
N Engl J Med 2000;342:365-73

Estimated Cumulative Rate of Appropriate Discharge



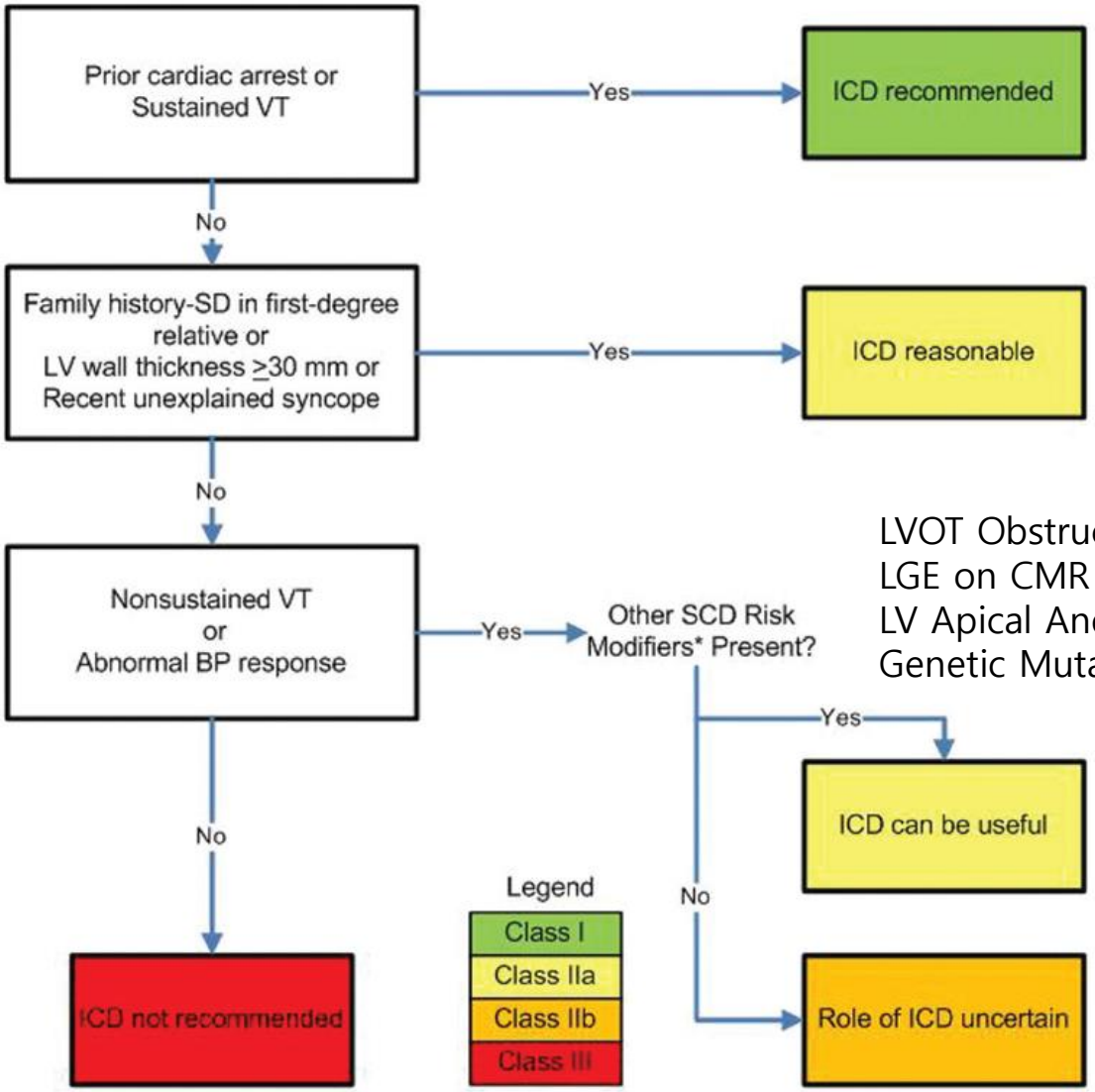
N Engl J Med 2000;342:365-73

Interval between implantation of ICD and First Discharge



N Engl J Med 2000;342:365-73

ICD Indication in Pt with HCM



LVOT Obstruction
LGE on CMR Imaging
LV Apical Aneurysm
Genetic Mutation

2011 ACCF/AHA guideline. Circulation 2011;124:e783-e831

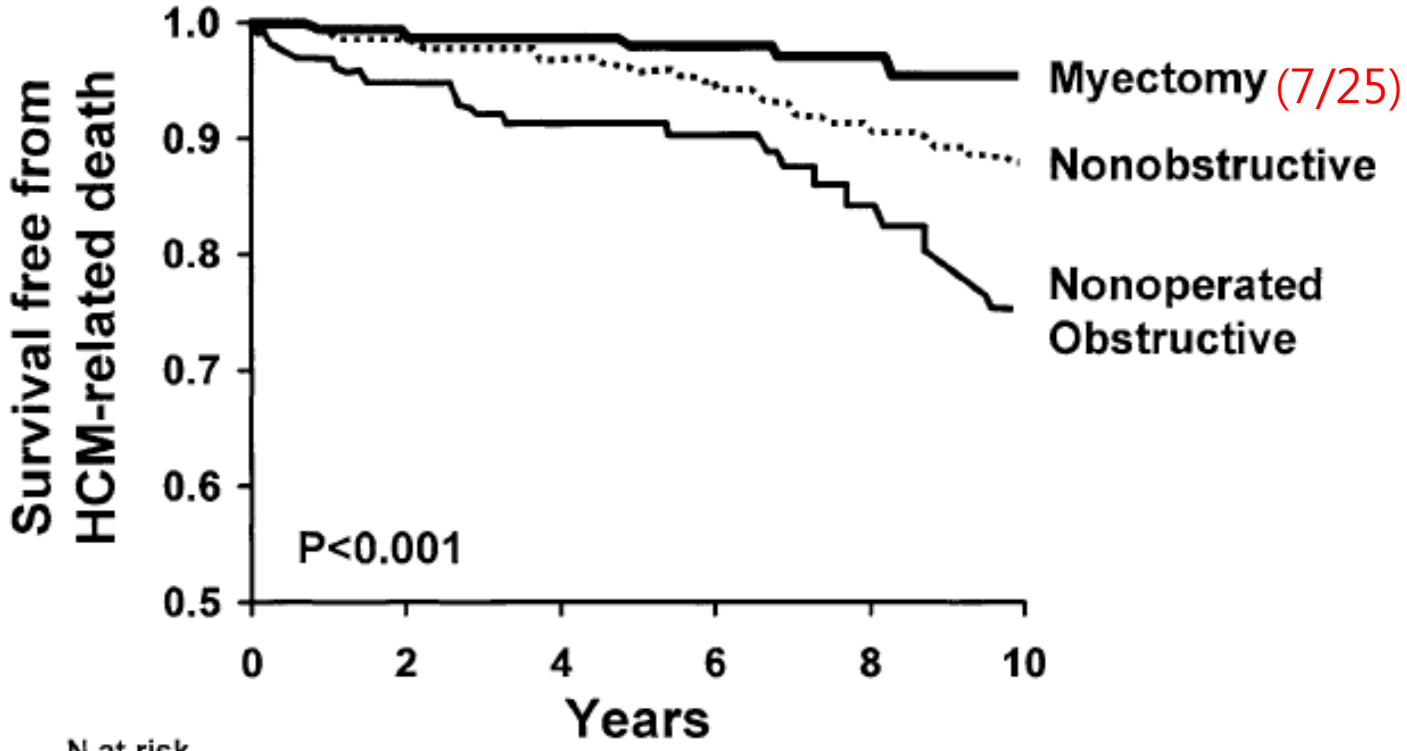
Conclusion

- There are many data which are strongly in favor of ICD implantation in high risk HCM.
- ICD provide highly effective discharges in primary prevention of SCD in HCM, significantly reduce mortality, improve long-term survival and increase quality-adjusted life expectancy.

Rebuttal

1. Long-Term Effect of Myectomy Remains Controversial

Long-Term Effect of Myectomy in HCM-1

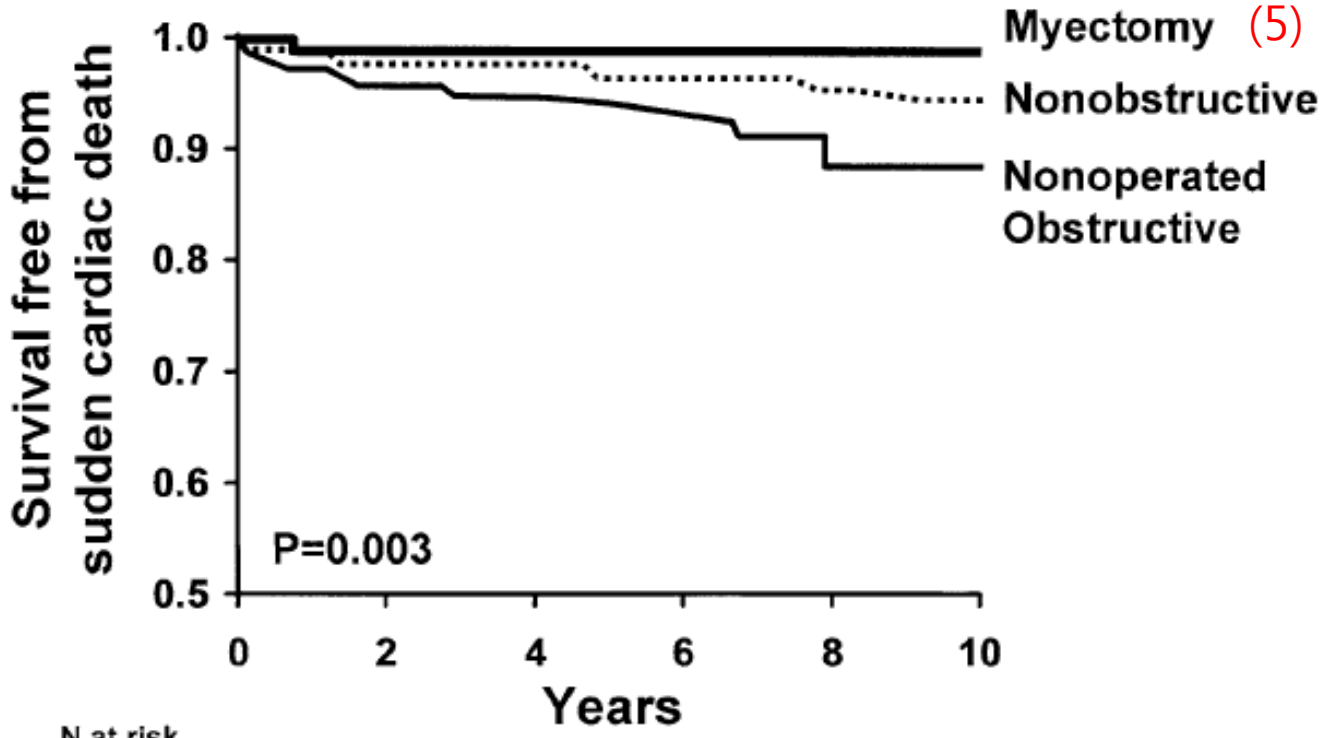


N at risk						
Myectomy	289	249	179	108	66	39
Nonobstructive	820	587	490	355	244	201
Nonoperated obstructive	228	146	106	69	42	28

Obstruction : LVOT Gradient ≥ 30mmHg under resting condtion

J Am Coll Cardiol 2005;46:470-6

Long-Term Effect of Myectomy in HCM-1



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Myectomy	289	249	179	108	66	39
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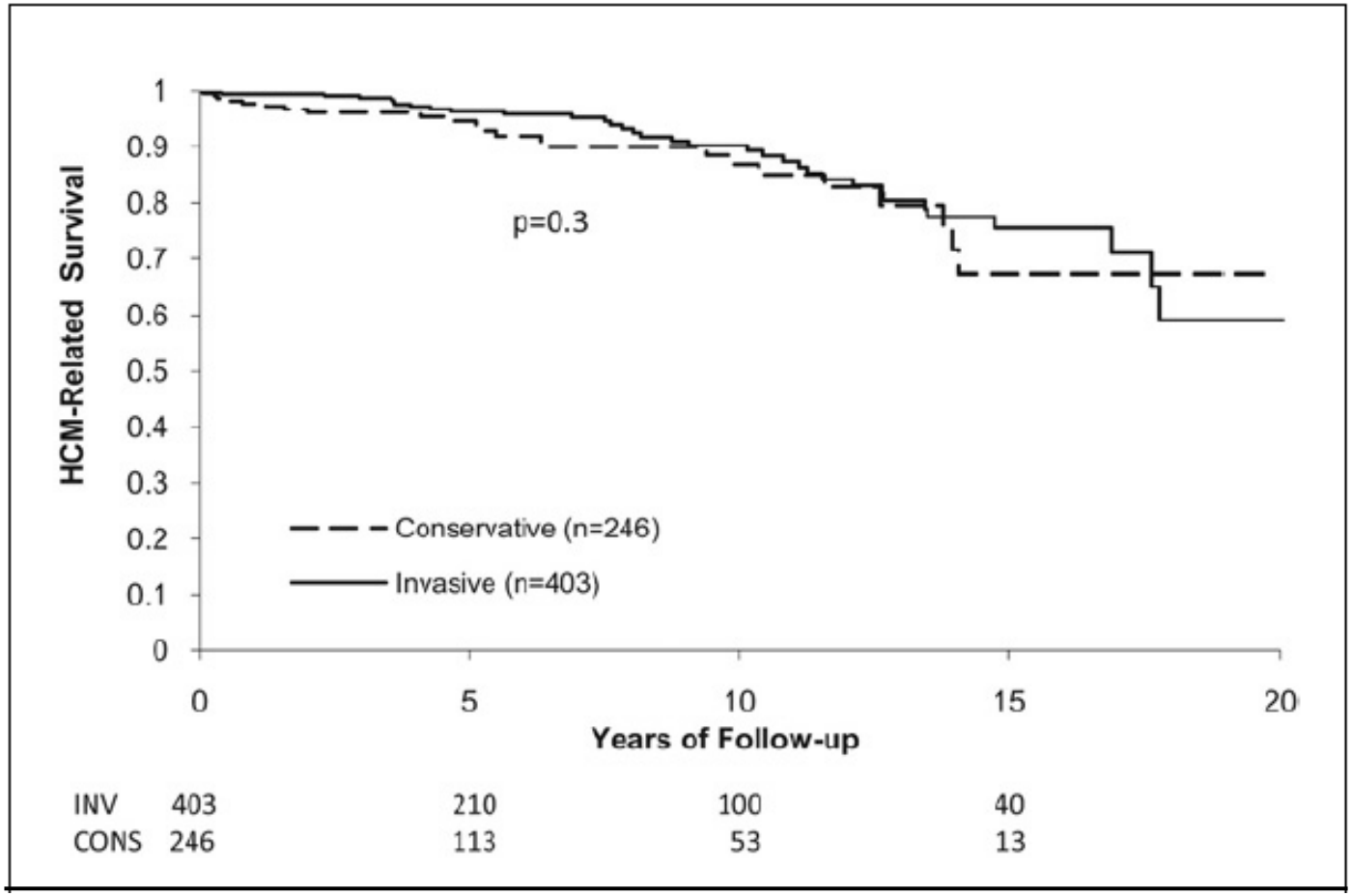
Nonoperated obstructive	228	146	106	69	42	28

J Am Coll Cardiol 2005;46:470-6

Long-Term Effect of Myectomy in HCM-2



Total of 55 patient(8.5%) received an ICD for primary or secondary prevention

J Am Coll Cardiol 2011;58:2313-21

Long-Term Effect of Myectomy in HCM-2

	Conservative Group (n = 246)	Invasive Group (n = 403)	Total (n = 649)
Mortality			
HCM-related mortality	19 (7.7)	28 (6.9)	47 (7.2)
Sudden cardiac death	8 (3.3)	7 (1.7)	15 (2.3)
Non–HCM-related death	16 (6.5)	8 (2.0)	24 (3.7)
Overall mortality	35 (14.2)	36 (8.9)	71 (10.9)
Equivalents of mortality			
Resuscitated cardiac arrest	2 (0.8)	4 (1.0)	6 (0.9)
Appropriate ICD discharge*	1 (0.4)	3 (0.7)	4 (0.6)
Total mortality and equivalents of mortality	38 (15.4)	43 (10.7)	81 (12.5)

Total of 55 patient(8.5%) received an ICD for primary or secondary prevention

J Am Coll Cardiol 2011;58:2313-21

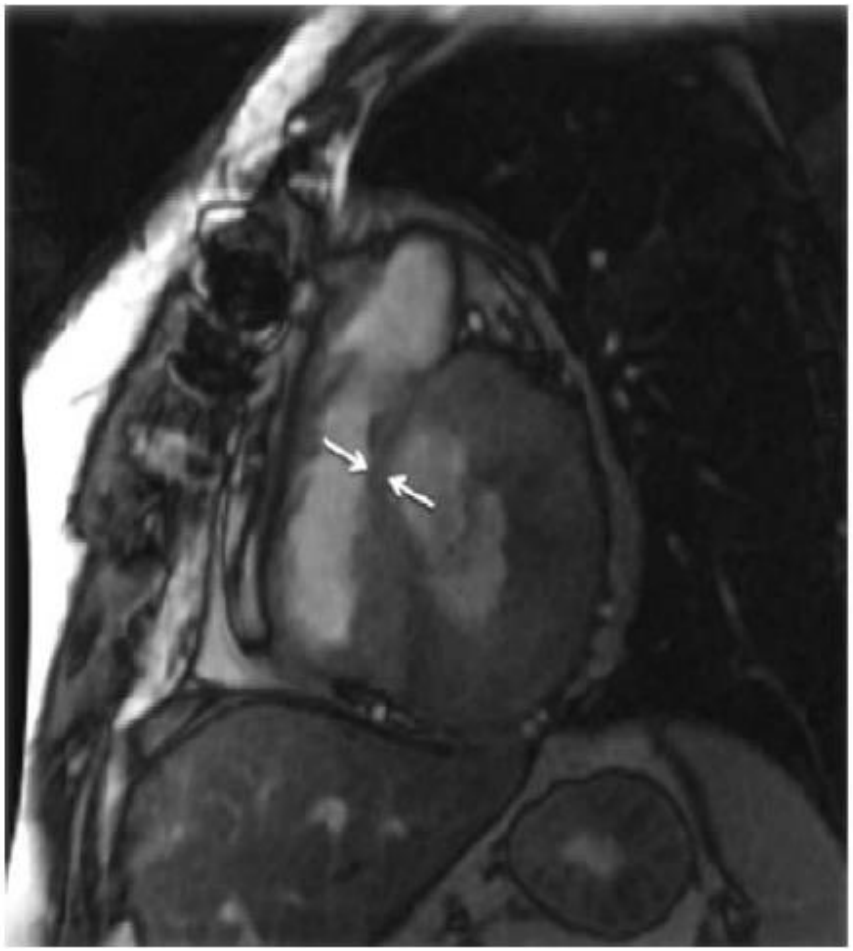
2. Complete Removal of Anatomical Substrate is Impossible.

Case Report

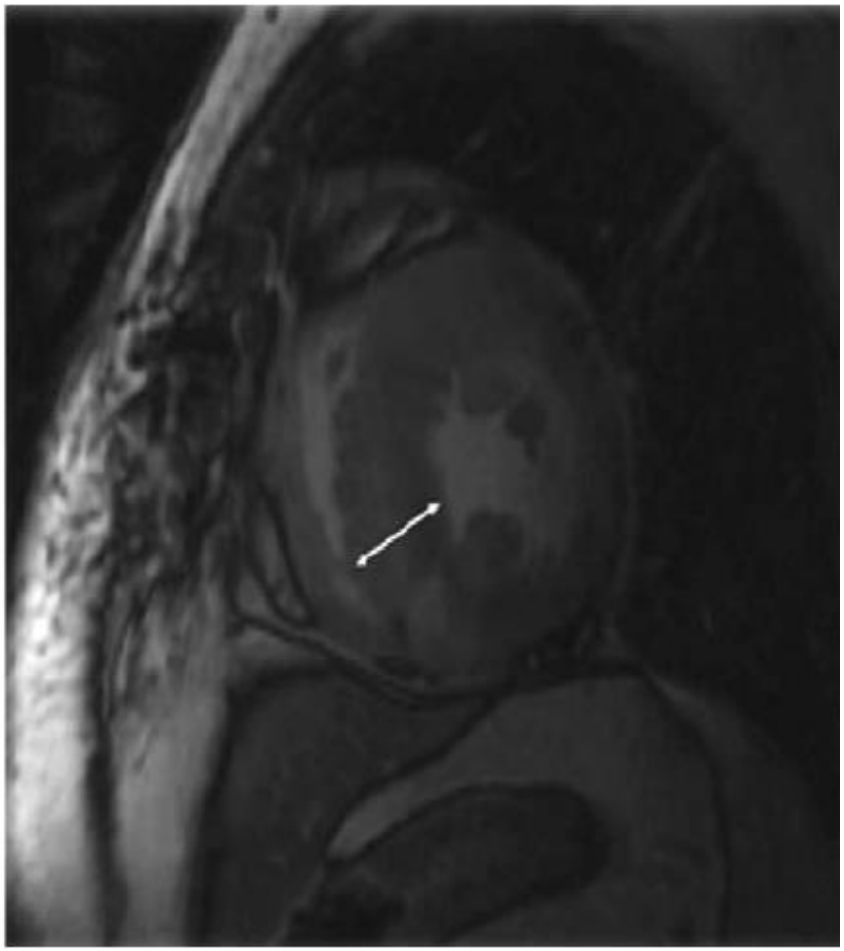
- 41/M(A), 45/M(B)
- Exertional dyspnea, chest tightness, palpitation, 1 episode of syncope
- FHx(-)
- TTE: maximal wall thickness 32mm
- LVOT gradient 143mmHg

Journal of Cardiology Case 2011;3:e65-e67

CMR after Septal Myectomy



Basel level(A)



Mid level(A)

Journal of Cardiology Case 2011;3:e65-e67

Natural Hx

- Case A ;

  ICD implantation 4 month after OP

  Appropriate shock, 6 month later

- Case B ;

  SCD 5 month after OP

3.The Recommendation for Septal Reduction Therapy is until limited

Recommendation-Invasive Tx

1. **Septal reduction therapy should be performed only by experienced operators* in the context of a comprehensive HCM clinical program and only for the treatment of eligible patients with severe drug-refractory symptoms and LVOT obstruction.†[272]** ***(Level of Evidence: C)***

   * Experienced operators are defined as an individual operator with a cumulative case volume of at least 20 procedures or an individual operator who is working in a dedicated HCM program with a cumulative total of at least 50 procedures (Section 6.2.2.3).

   † Eligible patients are defined by all of the following:

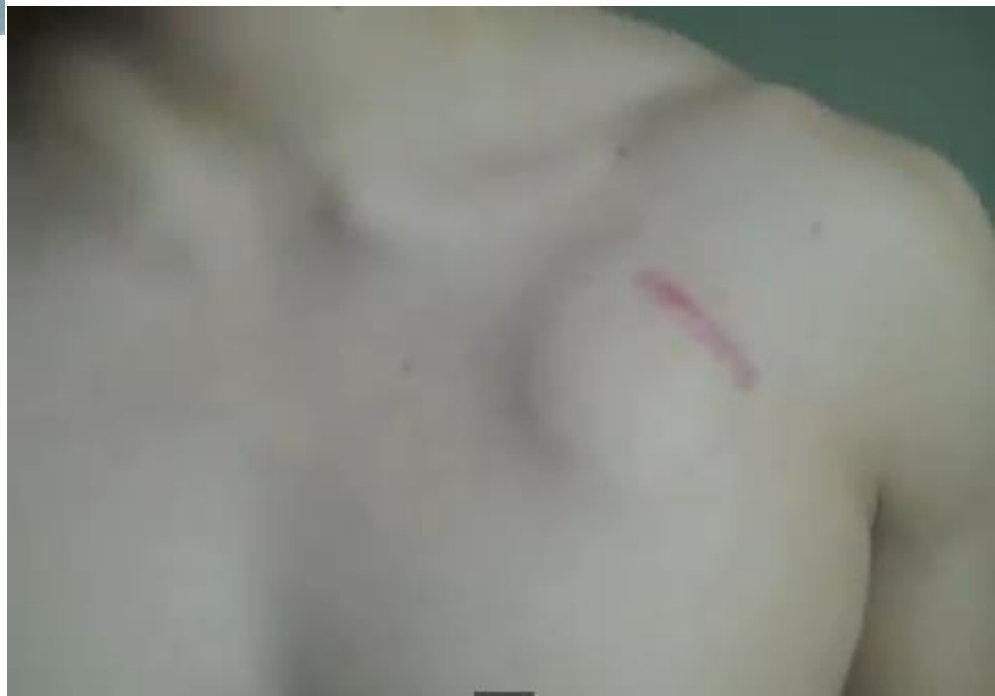
   a. Clinical: Severe dyspnea or chest pain (usually NYHA functional classes III or IV) or occasionally other exertional symptoms (such as syncope activity or quality of life despite optimal medical therapy.

   b. Hemodynamic: Dynamic LVOT gradient at rest or with physiologic provocation ≥50 mm Hg associated with septal hypertrophy and SAM of the mitral valve.

   c. Anatomic: Targeted anterior septal thickness sufficient to perform the procedure safely and effectively in the judgment of the individual operator.

2011 ACCF/AHA guideline. Circulation 2011;124:e783-e831

4. Patients and Physicians would prefer the less-invasive percutaneous procedure to open heart surgery



Complication

- Septal myectomy

  Overall risk of procedure highly depends on the experience of the operator

338 adult from 1978 to2002

Complications	n (%)
Early (within 30 days of myectomy)	
Postoperative death	5 (1.5)
Isolated myectomy group (n=249)	2 (0.8)
Myectomy and any concomitant surgical procedure(s) (n=89)	3 (3)
Permanent pacemaker	21 (6)
Ventricular septal defect	6 (2)
Early postoperative AF	102 (30)
Late (>30 days after myectomy)	
Subsequent surgical procedures	
Repeat myectomy	1 (0.3)
Ventricular septal defect repair	1 (0.3)
Mitral valve replacement	8 (2)
Pericardiectomy	1 (0.3)
Implantable cardioverter defibrillator	14 (4)
Cardiac transplantation	5 (1.5)
Serious cardiovascular events	
Congestive heart failure requiring hospitalization	44 (13)
Stroke	20 (6)
Arterial thromboembolic events	5 (1.5)
Cardiovascular cause of death	
Early postoperative death (during initial hospitalization for myectomy)	5 (1.5)
Sudden cardiac death	13 (4)
After myocardial infarction	2 (0.6)
Associated with congestive heart failure	15 (4)
After stroke	7 (2)
Early after cardiac transplantation	1 (0.3)
Total cardiovascular deaths	43 (13)

Circulatin.2005;111:2033-2041

Complication

- ICD implantation

  This procedure is ordinary procedure for general electrophysiologist

Complication	No of patients (%)
Pneumothorax (at ICD implant)	1 (<1)
Pericardial effusion (at ICD implant)	3 (2)
Pocket haematoma	4 (2)
Early (⩽1 month)	3
Late (with generator change)	1
Upper extremity deep venous thrombosis	1 (<1)
Lead revision	24 (13)
Acute (⩽24 h)	6
Chronic (>24 h)	18
ICD infection	8 (5)
Early (⩽1 month)	1
Late (>1 month)	7
ICD revision for high defibrillation threshold testing	6 (3)
Subcutaneous array	4
Lead revision	1
Generator change	1
Inappropriate shocks	42 (23)
Atrial fibrillation	20
Sinus tachycardia	16
Device malfunction	6

The procedure-related death is very rare.

Heart.2009;95:709-714

Conclusion

- ICD provide highly effective discharges in primary prevention of SCD in HCM.

- In high risk, obstructive HCM patient with drug refractory, severe Sx,

  Combination of Myectomy and ICD implantation may be benefit